Opercular syndrome in childhood with manic symptoms: a case report

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The Opercular Syndrome, also known as Foix-Chavany-Marie Syndrome, is characterized by malformation or lesion in the cortical perisylvian region (the most common being polymicrogyria, revealed by cranium magnetic resonance imaging - MRI). It displays several clinical manifestations such as epilepsy, motor deficits, pseudobulbar palsy, automatic-voluntary dissociation, speech difficulties, diplegia of masticatory muscles, and mental retardation. Although psychiatric symptoms are frequently cited, usually limited to unspecific psychopathological terms like "emotional lability", no detailed description of the psychiatric feature is available in the medical literature and rare report in children.

We report the case of a child with Opercular Syndrome who first presented with hyperactivity and euphoric mood symptoms, attended to a Child and Adolescence Affective Disorder Program, from nine to 18 years old.

Case

L, Caucasian, healthy girl, with family history of mood and substance use disorder. Since 3 years-old, due to psychomotor agitation, sleep disturbs and appetite complaints was treated with clonazepam, methylphenidate, sodium valproate, carbamazepine, oxcarbazepine, and risperidone, without any improvement. At 4 years-old, she also begam present prominent social inadequacy with disinhibition, learning disabilities, increasing impulsivity and aggressivity, which led to 3 different schools expelling her.

At 7years-old, she was first referred to our service, and call for attention the possibility of manic episode due to mood elation, pressure of speech, severe restlessness, decrease need for sleep, social inadequacy with disinhibited and hypersexualized behavior. There was no evidence of previous history of seizures, stroke, or depressive episodes. The clinical laboratory screening had normal results.

She initially received lithium carbonate 1050mg daily and risperidone 6 mg daily without any improvement. Still, no improvement when replaced lithium with sodium divalproate 1000 mg/day, and nor after replaced risperidone with olanzapine till 30 mg/day. The refractoriness of L. condition to drugs commonly used to treat mania has led to further investigation. The neurological assessment revealed pseudo-bulbar facie, mild drooling, craniofacial disproportion and left upper limb dysmetria, raising the diagnostic hypothesis of Opercular Syndrome. The cranium MRI showed posterior verticalization of sylvian fissures and sylvian fissure thickness, compatible with polymicrogyria thought Opercular Syndrome. The electroencephalography was normal.

Inpatient stay was then made necessary. Significant improvement was observed after reintroduction of lithium carbonate 750 mg/d (lithium dosage = 0.9 mEq/L) combined with olanzapine 30 mg/d. Despite mood stabilization, she kept on persistent hyperactivity symptoms pattern, then also use additional methylphenidate 10mg /3 times daily, later, switched to OROS methylphenidate 18mg once daily, with important hyperactivity improvement. With the same medications, L. eventually presented hyperthymic, however continued reasonably stable and remained in follow-up in our service until she was 18 years old.

Discussion

Several neurological disorders involving cortical and subcortical brain structures may present with behavior or emotional manifestation, which is difficult to distinguish from primary psychiatric disease, especially bipolar disorder

Our patient, due to the persistent euphoric mood, the rapid and abundant speech pattern and the decreased need for sleep, typical symptoms of mania, really made it difficult to visualize the primary neurological manifestations. Manic symptoms were reported as frequent after brain lesion; however, our patient has been monitored from nine to 18 y.o. which certainly guarantees that she has not suffered brain trauma, stroke, or tumor. Thus, the hypothesis of congenital opercular syndrome appears to be plausible.

The manifestation of emotional instability and psychotic presentations appears to be common in pediatric neurological syndrome, but psychiatrists tended to overlook neurological symptoms in psychiatric patients, and delay to uncover earlier symptoms of brain structure degenerative.

There is scarce literature regards psychopathological manifestations of Opercular Syndrome. The reported case highlights the need for more research on psychiatric symptoms related to this syndrome and its clinical management.

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